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ABSTRACT

Provided is a collection of presentations from a conference for ophthalmologists, optometrists, and educators meeting to exchange their concerns and share knowledge on various types of sensory handicaps of deaf blind children. The conference is seen as an introduction to the needs of deaf blind children and to the importance of stating diagnostic medical information in terms applicable to classroom training for vision specialists, and as an introduction to use of medical information in the development of classroom care and training for teachers. Included are the following presentations: "Usher's Syndrome: Problems and Some Solutions" (M. Vernon), "Ophthalmologic and Genetic Aspects of Deafness and Blindness" (D. Bergsma), "Examining the Deaf-Blind Child" (T. Johnston), "Assessing the Deaf-Blind Child" (L. Jenkins), and "Hints for Examining and Reporting the Vision of Deaf-Blind Children" (M. Efron). (IM)

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Proceedings

Workshop for Ophthalmologists, Optometrists, and Educators

Prepared under the direction of

William A. Blea, Project Director, Southwestern Region Deaf-Blind Center, California State Department of Education; and

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Preface

In 1975 the Southwestern Region Deaf-Blind Center sponsored a series of conferences in Los Angeles on sensory handicaps. At this particular conference, held September 11 and 12, noted ophthalmologists, optometrists, and educators from throughout the United States met and exchanged their concerns and shared their knowledge on the various types of sensory handicaps of deaf-blind children.

For some vision specialists, this conference was an introduction to the needs of the deaf-blind child and to the necessity of stating diagnostic medical information in terms that are applicable to vision training in the classroom. For some teachers, the conference was an introduction to the use of medical information in developing long-range care and training in the classroom and to methods for providing medical specialists with behavioral observations that may assist in more accurate diagnosis and assessment.

Only through the interaction between educators and specialists in all fields will the needs of the deaf-blind child be best met. We hope the papers presented at this conference will assist all of those concerned with the care and training of deaf-blind children in meeting the needs of their patients with increased effectiveness.

WILLIAM A. BLEA
Project Director
Southwestern Region Deaf-blind Center

Agenda for Workshop

Quality Inn Airport
5249 W. Century Boulevard
Los Angeles, California 90045

Thursday, September 11, 1975

8–9:30 a.m. ..... Registration

9:30–10:30 a.m. ..... Welcome

Center Concept I (Services for children, birth to age 21)

William A. Blea, Ed.D.
Project Director
Southwestern Region Deaf-Blind Center

Center Concept II (Services for deaf-blind, ages 21 and older)

Vera Schiller
Western Region Representative
National Center for Deaf-Blind Youth and Adults
Glendale, California

10:30–10:45 a.m. ..... Break

10:45–11:45 a.m.

McCay Vernon, Ph.D.
Department of Psychology
Western Maryland College
Westminster, Maryland

11:45 a.m.–12:15 p.m. ..... Group Discussion

12:15–1:30 p.m. ..... Lunch

1:30–2:30 p.m.

Donald Bergsma, M.D.
National Eye Institute
National Institute of Health
Bethesda, Maryland

2:30–3 p.m. ..... Discussion

Friday, September 12, 1975

9–10 a.m.

Ted L. Johnston, M.D.
Ophthalmologist
Cheyenne Eye Clinic
Cheyenne, Wyoming

10–10:30 a.m. ..... Discussion

10:30–10:45 a.m. ..... Break

10:45–11:45 a.m.

Laurice Jenkins, Evaluation Center Tape
Educational Assessment Center
California School for the Blind
Berkeley, California

11:45 a.m.–1 p.m. ..... Lunch

1–2 p.m.

Marvin Efron, O.D., Ph.D.
Optometrist
West Columbia, South Carolina

2–2:30 p.m. ..... Discussion

2:30 p.m. ..... Workshop Summary

Contents

	Page
Preface	iii
Agenda for Workshop	iv
Usher's Syndrome: Problems and Some Solutions	1
Patient Case Histories	1
Fate of Victims	2
Problems with Delivery of Services	3
Solutions to Problems	3
Summary	6
Ophthalmologic and Genetic Aspects of Deafness and Blindness	9
Definition of Blindness	9
Causes of Visual Loss	9
Genetic Considerations	12
Patient Counseling	12
Rehabilitation and Screening Programs	12
Examining the Deaf-Blind Child	15
Examination Location	16
Eye Screening Evaluations	16
Examination Instruments	17
Conclusions	17
Assessing the Deaf-Blind Child	19
Enrollment Procedure	19
Evaluation Period	19
Entrance Criteria	20
Visual Acuity Assessment	20
Hints for Examining and Reporting the Vision of Deaf-Blind Children	23
Examination Conditions	23
Measurement of Visual Acuity	24
Determining Level of Function	24
Examination Report	24

Usher's Syndrome: Problems and Some Solutions

Presented by McCay Vernon
Department of Psychology, Western Maryland College

Usher's syndrome is the leading cause of deafness and blindness in the most scientifically advanced countries throughout the world (Vernon, 1974). The disease involves primarily a profound congenital loss of hearing and a progressive loss of vision due to retinitis pigmentosa. It is transmitted genetically by an autosomal recessive gene.

Ironically, most ophthalmologists, otolaryngologists, optometrists, psychologists, pediatricians, and other professionals upon whom patients with Usher's syndrome depend for diagnosis and treatment are not even aware of the disease. Because of this lack of knowledge about the disease, little has been done to treat, prevent, or understand Usher's syndrome or the needs of its victims.

Professionals are not familiar with Usher's syndrome for a number of reasons. The major reason is that those who work with the disease have failed to communicate the necessary information to those in the field. Another reason is that Usher's syndrome is a rare condition which occurs in about 3 out of every 100,000 persons (Vernon, 1969). However, among certain high-risk populations this condition is not rare at all. For example, 3 to 6 percent of those born deaf have Usher's syndrome (Bergsma, 1973). Among those with retinitis pigmentosa, 4.3 to 6.3 percent have a hearing loss (McLeod and others, 1971), which by definition means that they have the disease.

Ophthalmologists read about retinitis pigmentosa in the trade journals, and they diagnose the disease. However, they are often unfamiliar with the genetic aspects of the disease and the relationship of deafness to the visual problem. The otologist diagnoses neural deafness but does not associate it with the visual problem or with the hereditary etiology. The psychologist usually does not understand the visual, auditory, or genetic aspects of the disease and does not communicate well enough with the deaf patient to obtain much valid psychological information. Therefore, the problems of the patient with Usher's syndrome are treated in a fragmented way, which is grossly inadequate and inappropriate. The examples cited in the following sections will illustrate some of these problems.

Patient Case Histories

The first case involves a twenty-six-year-old deaf man who had attended an excellent school for the deaf. The school had a full-time psychologist, an otolaryngologist, an ophthalmologist, and an optometrist, all of whom examined students on an annual basis. Despite having clear evidence of night blindness, tunnel vision, and vertigo, the man graduated from the school at the age of twenty without having his case of Usher's syndrome diagnosed. He subsequently had physical and otological examinations at the Division of Vocational Rehabilitation. Although these tests did detect that he had neural deafness, they did not reveal that he had Usher's syndrome. Later, after he had been in a series of automobile accidents, the man submitted himself to psychological evaluation, which revealed that he had a greatly reduced field of vision. He was then referred to an ophthalmologist who was thoroughly familiar with Usher's syndrome. The ophthalmologist discovered that he had only a 17-degree field of vision remaining. This man had been endangering his life and the lives of others while driving a car and working at a hazardous job in a meat plant because he did not know that he was legally blind. His case had not been detected by specialists in schools for the deaf, where 3 to 6 percent of the students have the disease.

The second example involves another young man who graduated from a college for the deaf. He subsequently taught for three years in a school for deaf children. A layperson at the school who happened to be familiar with Usher's syndrome

referred him to an ophthalmologist who discovered that he had the disease. This patient subsequently went back to his college and identified five fellow students who had Usher's syndrome. Although all of the students had been examined by medical and other professionals at the college and in schools for the deaf, none had been told that he or she had the disease.

One final example involves a highly successful, retired, sixty-year-old State Department official who had been treated for years for retinitis pigmentosa. This man was never told that he had Usher's syndrome because none of those who treated him were familiar with the disease. Finally, late in life, he happened to read an article on Usher's syndrome. From the article he diagnosed that he had the disease. His immediate fear was that his children and grandchildren might also have Usher's syndrome.

These cases point up the fact that, until the professionals who see patients with Usher's syndrome are made aware of the disease, many cases will go undetected beyond the time that they are easily diagnosed. This ignorance among the professionals who regularly treat deaf persons is especially unfortunate because the deaf are the high-risk group for the disease.

So far little has been done to get information to physicians, to people in schools where deaf children are taught, to speech and hearing clinics, to vocational rehabilitation counselors, or to similar groups that should be referring these patients for ophthalmological evaluation. Consequently, individuals with the disease are making vocational and professional plans without knowing that they will eventually be blind. They get married and have children, unaware of their impending blindness and inability to support their families. Many of them drive cars and work in dangerous occupations, oblivious to their visual problem or its implications.

The time has arrived to correct this problem. The cost would be small in comparison to the human misery now being created. In fact, an active prevention program that includes genetic screening and counseling would yield significant financial savings.

Fate of Victims

All persons with Usher's syndrome are born either deaf or profoundly hard of hearing. They are usually brought to one of the approximately 700 speech and hearing clinics in this country during their preschool years (Craig, 1975). Thus, we have an early centralization of the high-risk population with Usher's syndrome. A primary target population for education about the disease should be the professionals who serve in these clinics. A few cases of Usher's syndrome are clearly diagnosable during preschool years, but they are the exception.

Following speech and hearing therapy or preschool education, most children with Usher's syndrome are sent to a school for the deaf or to a special education program for the deaf and hard of hearing. The high-risk population with Usher's syndrome is again highly centralized and readily available for diagnosis and rehabilitation. In addition, the professionals who need an education about Usher's syndrome are also centralized and available.

During the school years, the symptom of night blindness inevitably appears and is easily identified as a major problem. Often, in residential schools for the deaf, one will see several teenage deaf students being helped around at night because of their night blindness.

The visual field also begins to narrow (tunnel vision) during this period, especially in the latter teenage years. Somewhere between the age of eight and the teens screening for Usher's syndrome becomes a highly feasible, effective, relatively simple, and not unduly expensive procedure (Bergsma, 1973). However, at present, no school or college for the deaf has such a screening program. Consequently, the average person with Usher's syndrome leaves school or college unaware that he or she has the disease. Usually, the person has been prepared by the school for the deaf for a job that emphasizes his or her visual skills.

Shortly after graduating from school most persons with Usher's syndrome get married and start a family. The spouse is usually deaf, quite often due to genetic causes, which increases the probability of the couple's offspring having Usher's syndrome. Typically, the person's vision starts to interfere with his or her daily activities, especially driving, in the late twenties and early thirties. Central vision acuity usually remains within normal limits, although cataracts may appear.

Most victims of Usher's syndrome usually have their case diagnosed by an ophthalmologist as retinitis pigmentosa when they are between twenty and forty years of age. Very often, however, they are not told of this fact nor are they informed of the prognosis for their vision or the genetic nature of their problem. The traumatic task of telling a person that he or she is going to be deaf and blind is one that many human beings do not care to undertake, including the ophthalmologists. When

and how the diagnosis should be presented to the victim is a profound psychomedical issue.

In time persons with Usher's syndrome have to stop driving. They are forced to give up their jobs. Gradually, their deaf friends start avoiding them because it is difficult for them to communicate in sign language. They cannot read signs unless people spell directly into their face or hand, or unless they stand in the light. They are hard to contact because they are unable to drive or use the telephone. They are also a burden because they need help to walk at night.

Consequently, most persons with Usher's syndrome find themselves unemployed and socially isolated some time between their late twenties and forties. Frequently, divorce occurs. By this time the children usually have left home. The victim is left with almost no human or financial resources. After being discouraged from social interaction with hearing people from childhood because of their deafness, these people now find that deaf people reject them because of their blindness. In addition, blind people do not wish to interact with them because the blind depend on speech and hearing and cannot communicate in sign language.

Despite the fact that they are neither retarded nor psychotic, many persons with Usher's syndrome are placed in state hospitals for the mentally ill or the mentally retarded in their later years (Bettica, undated). Often, the reason for putting them into such hospitals is that there is no other place for them.

One of the ironies of the tragic neglect of persons with Usher's syndrome is that, although the disease is the leading cause of deafness and blindness, the victims of rubella syndrome have received the most attention (Brewer and Kakalik, 1974). In fact the regional Centers for Deaf-Blind Children and the National Center for Deaf-Blind Youths and Adults are mainly an outgrowth of rubella syndrome rather than Usher's syndrome (Brewer and Kakalik, 1974).

Problems with Delivery of Services

The major problem with the delivery of services is that there are not enough services to deliver. Secondly, the few services that are available are so fragmented that it is difficult for persons with Usher's syndrome to understand, coordinate, and utilize them (Brewer and Kakalik, 1974).

Poor Prevention Services

Because Usher's syndrome is not diagnosed by professionals, prevention is minimal. Yet, because the disease is a genetic, autosomal, and recessive disorder, some prevention is possible through genetic counseling (Nance, 1973). For example, if the disease were diagnosed early in the child's life, the parents could be advised of the probabilities of having additional children with Usher's syndrome. In my own work I have contact with several families that have four or more children with this disease. If these parents had been informed of the genetic aspects of their disorder and the probability of it being passed on to their children, they could have taken preventive measures.

If techniques were developed for identifying carriers of Usher's syndrome, major inroads for prevention of the disease through genetic screening and counseling would be possible. Similarly, options such as therapeutic abortion would become a reality if the disease, which is believed to be caused by an inborn error of metabolism, could be diagnosed biochemically in the fetus of the carrier.

Visual screening is another possibility. For example, we now have mandatory screening of all infants for phenylketonuria (PKU), which affects only 1 out of 10,000 children. Screening of the approximately 10,000 high school age deaf students for Usher's syndrome under the plan developed by Bergsma (1973) would provide a 3 to 6 percent yield as compared to the tenth of one percent yield provided by PKU screening.

Lack of Effective Planning

Case finding among the deaf-blind is notoriously poor, especially among people with Usher's syndrome. Consequently, there is no basis for determining how many people have the disease. A combination of figures from the National Census of the Deaf (Schein and Delk, 1975) and Bergsma's prevalence data (Bergsma, 1973) indicate that there are approximately 16,000 persons in the United States with Usher's syndrome. However, it is doubtful whether 1,000 of these cases have been identified. Obviously, effective planning is impossible with such poor case finding.

Solutions to Problems

One can point out the inadequate, inhumane, and fragmented nature of the existing treatment of persons with Usher's syndrome, but it is quite another matter to operationally spell out solutions

to the problem. However, we will attempt to do this in the remainder of this discussion.

Prevention

Because Usher's syndrome is incurable and involves the tragic double handicap of deafness and blindness, primary efforts should be devoted to its prevention.

Screening and genetic counseling. The best approach to the prevention of Usher's syndrome is through visual screening and genetic counseling. As indicated earlier the high-risk population for the disease is the deaf. During school years and to a certain extent in later life deaf people are a highly centralized and available population. Furthermore, the yield of visual screening of the deaf would be around 4 percent (Bergsma, 1973), which would increase to 8 percent if only the genetically deaf were screened (Vernon, 1969). Bergsma (1973) has developed an inexpensive and practical pilot approach for doing this. The cost of Bergsma's proposed screening is infinitesimal as compared to the cost of educating, training, or just providing custodial care for a deaf-blind person. For example, to educate one deaf-blind child for 13 years costs approximately $182,000 (Brewer and Kakalik, 1974). This amount includes the cost of the screening program for several years.

If screening were combined with follow-up genetic counseling, Usher's syndrome still would not be eradicated because it is an autosomal recessive disease (Nance, 1973). However, by counseling the patient with the disease, the patient's parents, and the relatives, the number of cases of Usher's syndrome could be reduced significantly.

Screening and genetic counseling pose many ethical and philosophical problems, which is one reason Bergsma has advocated only a pilot program initially. This may also be why he has revised the age at which screening should be conducted from eight years of age to sometime during the teen years (Bergsma, 1973).

Research. Evidence from past research indicates that carriers of Usher's syndrome can be detected by audiological or possibly biochemical testing (Kloepfer, Laguaite, and McLaurin, 1966; McLeod and others, 1971; Vernon, 1969). Further research on carrier detection would have a high probability of success and would not be unduly expensive. Once a technique is developed to identify carriers, Usher's syndrome, like any other genetic disorder, will become much more amenable to prevention through screening and genetic counseling.

In addition to giving hope for prevention, research also holds forth reasonable expectations for the cure or arrest of the disease. Usher's syndrome is believed to be caused by an inborn metabolism error (Vernon, 1973). A similar condition, which also causes deafness and blindness, Refsum's syndrome, has been attributed to an enzymatic defect that can be treated with a dietary supplement (Vernon, 1974). If research revealed the suspected biochemical defect that causes Usher's syndrome, the deficiency might be supplemented, thereby arresting, preventing, or at least reducing the effects of the disease (Vernon, 1974).

None of the proposed research is excessively expensive. All of it is cost effective in that the expenditures do not approach the cost of maintaining even minimal services for deaf-blind people (Brewer and Kakalik, 1974).

Public education. Little or no research, diagnosis, screening, genetic counseling, prevention, rehabilitation, or treatment will be done until physicians, optometrists, and other professionals who work with deaf children know about Usher's syndrome. Therefore, the most urgent priority has to be the communication of information about Usher's syndrome to professionals. Thus far embryonic efforts have been made. For example, the National Foundation for the March of Dimes has circulated an article on Usher's syndrome to geneticists, Gallaudet College has held two symposiums on the disease and published the proceedings, and the Southwestern Region Deaf-Blind Center has hosted this workshop and has published the proceedings.

Another possibility is the production of three documentary films on Usher's syndrome, such as the following:

1. The first film would be for ophthalmologists, geneticists, optometrists, researchers, and others familiar with medicine or vision. For example, if ophthalmologists knew about Usher's syndrome, they would routinely check all deaf patients by at least examining the periphery of the retina with their ophthalmoscope, preferably after dilating the pupil (Bergsma, 1973). Visual field testing would be done more often, and referrals would be made for dark adaptation testing.

2. The second film would be for teachers, rehabilitation counselors, audiologists, speech pathologists, and others whose work involves frequent contact with deaf or hard of hearing children. If these individuals were familiarized with Usher's syndrome, they could very easily identify

the early symptoms, such as night blindness and tunnel vision, and refer the patients to ophthalmologists.

3. The third film would be for the lay public, including service clubs. For example, Lions International has blindness as its primary interest and deafness as its secondary interest. Usher's syndrome combines these two conditions and would be an ideal project for the Lions. With their resources they could quickly and easily raise the funds needed in work for preventing Usher's syndrome and for sponsoring the research to develop a treatment. This third film for the lay public would also be very useful on public television in creating national awareness of the problem of Usher's syndrome.

The need to inform physicians is greater today than ever before because of a fundamental problem faced by doctors. In the past a physician spent a great deal of time treating patients who had one of a relatively small number of infectious diseases. Thus, by learning to diagnose a moderate number of these diseases, the doctor was able to diagnose the illnesses of a high percent of his or her patients. With the advent of antibiotics, the diagnosis and treatment of patients with infectious diseases takes a relatively small amount of a doctor's time. In fact, these diseases are often treated and cured without the physician knowing the organism that caused them. Consequently, the doctor is left with a disproportionate number of his or her patients who have one of some 2,000 known rare genetic diseases. Today the doctor is expected to be able to diagnose rare genetic diseases that he or she may see only once or twice during an entire career in medicine. Doctors should not be expected to do this if they are not provided with the necessary information about these diseases. Films, articles, audiotapes, and television are ways of providing this information.

Rehabilitation and Services

While the ultimate solution of the problems of Usher's syndrome is prevention or cure, we must face the fact that an estimated 16,000 people now have the disease. What should be done for these individuals?

Centralization of services. The presently fragmented, inadequate, and wasteful services need to be centralized (Brewer and Kakalik, 1974). Servicing a population of 16,000 individuals who have specialized, expensive needs is difficult when they are scattered throughout 50 separate state programs. A federal program exists in the Bureau of Education for the Handicapped (BEH); however, it serves primarily the 4,000 to 5,000 rubella deaf-blind children.

Today everyone expects the federal government to solve all of his or her problems, often without adequate justification. However, facts make it clear that low incidence disorders such as Usher's syndrome must be handled at the federal level. Perhaps the National Center for Deaf-Blind Youths and Adults or the Regional Centers of the Bureau of Education for the Handicapped are the mechanisms. The point is that a centralized program is needed for the deaf and blind, including those with Usher's syndrome. Such a program is a necessary first step in providing appropriate services.

Deaf-Blind Regional Centers. A need exists for actual service, not just diagnosis and evaluation. In the field of rehabilitation, everyone wants to evaluate and diagnose, but few wish to assume the responsibility for the patient on a day-to-day basis. The easy way out is just to label patients or describe their assets and liabilities and to forget about doing something for them. This is a very real problem for the Regional Centers, which are primarily diagnostic facilities (Dantona, 1973).

Direct service must be made available for deaf-blind people. My own view is that such services should be two dimensional.

1. First, one or two facilities should be available in the United States to which deaf-blind adults having IQs at the retarded level could go and live. Those who are able to work could have jobs, while those incapable of working at a job would be supported by the facility. The creation of two such centers would ensure that the deaf-blind would not have to face the social isolation neglect, and abuse that now confronts most of these people. Many deaf-blind persons who now come to the National Center express a strong desire for an option of this kind.

2. A second dimension to the service program would be to offer deaf-blind persons who prefer to remain in their own community the necessary supportive services. For example, many persons with Usher's syndrome have gone to the National Center for Deaf-Blind Youths and Adults in New York and developed self-help and/or vocational skills that enabled them to return to their hometowns and live with their families. This option should be opened to all for whom it is realistic and who seek it.

Available Services

A listing of some of the major services available to all blind persons, including those with Usher's syndrome, is as follows:

1. *The National Center for Deaf-Blind Youths and Adults, 105 Fifth Avenue, New Hyde Park, NY 11040.* The National Center offers diagnosis, evaluation, training, and follow-up to any deaf-blind person meeting the admission requirements. The National Center should be one of the first places contacted by any deaf-blind person or his or her family.
2. *Bureau of Education for the Handicapped (BEH), Department of Health, Education, and Welfare, Washington, DC 20201.* Almost every community has a vocational rehabilitation counselor. He or she can provide a deaf-blind person with medical, educational, and other services, provided the deaf-blind person meets certain standards and provided the counselor's funds are adequate. Ironically, in these times of high unemployment, the money for providing job training and education for handicapped people has been drastically cut. Furthermore, many of the rehabilitation counselors not only cannot communicate with deaf-blind people but also have a limited understanding of their potential, their needs, and what is available for them.
4. *Supplementary Security Income (SSI), Social Security Administration, Department of Health, Education, and Welfare, Washington, DC 20201.* This is a new government program which offers many deaf-blind people a regular subsistence income. Enrollment in SSI also makes training money available from the Division of Vocational Rehabilitation that would otherwise be frozen (Vernon, 1975). Almost every community has an SSI office.
5. *The Committee for the Deaf-Blind, American Council for Blind, Inc., 539 New England Building, Topeka, KS 66603.* Formed in 1973, this committee is a division of the American Council of the Blind. It provides advice and some help to deaf-blind people. Half of its members are deaf-blind.
6. *National Retinitis Pigmentosa Foundation, Rolling Park Building, 8331 Mindale Circle, Baltimore, MD 21207.* The foundation conducts research on retinitis pigmentosa and hopes to discover a cure. It also keeps a national registry of people with Usher's syndrome, publishes information on devices to help deaf-blind people, and reports on the latest research findings on retinitis pigmentosa.
7. *Public Service Program of Gallaudet College, Gallaudet College, Kendall Green, Washington, DC 20002.* This program has sponsored two symposiums on Usher's syndrome and published information about the disease. The program is new, and the college has not spelled out what other services will be offered.
8. *National Association of the Deaf (NAD), 814 Thayer Avenue, Silver Spring, MD 20910.* The NAD provides some literature on Usher's syndrome and has a section on deaf-blindness. A small but significant percent of their membership have Usher's syndrome. As the world's largest organization of the deaf, they have the best contact with people with Usher's syndrome.
9. *International Association of Parents of the Deaf, 814 Thayer Avenue, Silver Spring, MD 20910.* This group has just established a committee for parents of deaf-blind children. Their services are only beginning, but they could develop into a major asset for a family who has just discovered that their child has Usher's syndrome or is deaf-blind for other reasons.

Summary

I have attempted to provide an overview of Usher's syndrome by touching on the nature of the disease, by describing the impact the disease has on the victim, by presenting the state-of-the-knowledge on the disease, and by discussing the services available for patients. Appropriate programs for prevention of the disease, research on the disease, and services for individuals with the disease have been described in operational terms.

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Ophthalmologic and Genetic Aspects of Deafness and Blindness

Presented by Donald R. Bergsma, M.D.,
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I work with patients who have retinitis pigmentosa and other forms of retina degeneration. Among these patients are a number of people who are deaf or have Usher's syndrome, which is a major cause of deafness and blindness. However, I would like to point out that Usher's syndrome is only one of more than 100 causes of deafness and blindness. Therefore, I am going to discuss briefly some of the various causes of deafness and blindness and then cover in detail the work that is being done on retinitis pigmentosa and Usher's syndrome. I hope that you will be able to apply some of this information to the cases you may encounter in your practices.

First, I think everyone should realize that being deaf is no protection against becoming blind. Conversely, being blind is no protection against becoming deaf. The incidence of deafness is higher among the blind than it is among the general population, while the incidence of blindness is higher among the deaf than it is among the general population.

In addition, early diagnosis and screening is more important for some of the other causes of deafness and blindness than it is for Usher's syndrome. For instance, the progression of glaucoma can be slowed down considerably with early treatment; therefore, one should attempt to detect the treatable or the preventable cases. Screening programs should be designed to detect a variety of diseases rather than just one disease.

Definition of Blindness

Before discussing blindness, we should define it because very few individuals are totally blind or have no light perception in either eye. The most common definition of legal blindness is 20/200 vision or worse in the better eye with the best refracted correction, or less than 20 degrees of horizontal field in the better eye. A person with normal sight can see an object at 200 feet, whereas the handicapped person can only see the object at 20 feet. These figures are measurements of visual acuity or central vision, which involve such functions as reading, driving, and observing detail.

The remainder of the definition of legal blindness—less than 20 degrees of horizontal visual field—refers to side vision. This definition is incomplete because the degree to which one can see to the side depends on the size and the brightness of the object, the contrast between the object and the background, and the speed at which the object is moving. The side vision measurement is extremely important because it involves one's mobility and general observation skills. Knowing where to look and where to use central vision depends to some extent on peripheral vision. I consider patients with severe loss of side vision to be more handicapped than patients with severe loss of central vision. A person with macular degeneration (loss of central vision) is not able to drive, but he or she can read by holding the material close or by using special lenses. This patient is classified as legally blind but has functional vision, except for driving. On the other hand, a person with good central vision that is limited to 2 or 3 degrees is handicapped for mobility and for reading. Children in particular do surprisingly well in reading even though they have poor distance vision. Therefore, a person is not totally handicapped just because he or she is legally blind or has 20/200 vision.

Causes of Visual Loss

The degree of blindness also depends on whether visual loss is stable or incurable, whether it involves one eye or both eyes, whether it is a partial or a complete loss, and whether it is a part of a syndrome. All of these factors will vary considerably with the cause. A brief description of some of the major causes of visual loss is presented in the following sections.

Cataracts

Perhaps the most familiar cause of visual loss is the cataract. The term *cataract* is defined as a clouding of the lens of the eye or its capsule, which obstructs the passage of light. The term *opacity* is used in referring to cataracts to avoid scaring the patient. As people grow older, they usually develop some minor degree of opacity. If the doctor uses the word cataract, the patient assumes that he or she is going blind and that he or she needs a lengthly operation; therefore, the term *lens opacity* is frequently used.

Cataracts can occur at all ages. For example, people with diabetes mellitus usually get cataracts in late adult life, while inherited cataracts usually occur very early in life. When cataracts are severe, the main therapy is surgical removal. Rubella babies frequently have cataracts along with an inflammation that makes an operation more difficult. On the other hand, patients with Usher's syndrome and retinitis pigmentosa often have cataracts, but the cataracts are usually very mild and progress slowly.

Neurological Diseases

Neurological diseases also cause blindness or visual impairment. Efferent neurological diseases are diseases of the nerves that run from the brain to the eye. These nerves control the eye muscles used for eye movement, lid closure, pupillary reaction, and so forth. Strabismus is a subcategory in which eye movements are not coordinated properly, and the two eyes do not line up visually. However, strabismus occurs many times as an isolated finding; otherwise, the person is neurologically normal.

Miscellaneous Diseases and Infections

The most common tumor in childhood is retinoblastoma. About 30 percent of these tumors are genetic, and the other 70 percent are spontaneous. I do not know of any particular association between tumors and deafness, but just because one is deaf does not mean that he or she can not get a tumor.

The incidence of early deafness is higher among diabetics probably because of the early onset of arteriosclerosis of the small blood vessels to the inner ear. People who have diabetes in early to late adulthood often get retinal complications, including bleeding, scarring of the retina, and retinal detachments. These conditions are sometimes combined with the early onset of deafness.

Sickle cell anemia is a genetic disease fairly common among blacks. The disease is relatively uncommon because it is recessive. One has to inherit the trait from both parents to get the disease, which causes retinal changes similar to those in diabetes. The sickle cell tends to cause clotting in the smaller blood vessels throughout the body.

Hypertension affects the human body in many ways. This condition contributes to the early onset of hearing loss and retinal damage and should be checked.

The incidence of retrolental fibroplasia was very high when oxygen therapy was first used in the 1940s and early 1950s. Infants who were born prematurely developed scar tissue behind the lens of the eye because of the high oxygen level required to keep them alive.

Finally, infectious diseases can cause loss of vision after a trauma. This possibility is associated with a variety of diseases.

Retinal Disease

Retinal disease is the largest category of diseases that cause loss of vision. About 40 percent of blindness is caused by retinal disease. At the present time we do not have cures for most of the retinal diseases.

Usher's syndrome is, by definition, a combination of deafness and retinitis pigmentosa. Some patients with only a partial loss of hearing develop retinitis pigmentosa, and others, the majority in fact, who have severe congenital deafness, develop the disease during the teenage years. This disease is a slow, progressive degeneration of the retina and is characterized by the loss of side and night vision, the development of pigmentary clumping, and the loss of electrical response.

Retinitis pigmentosa symptoms. The first symptom of typical retinitis pigmentosa is night blindness. The patient takes longer to adapt to the dark. He or she often does very well in daylight or good room light, but he or she can become completely lost in the dark.

Another typical symptom is poor side vision, which is tested in terms of visual field loss. This condition is usually detectable by the teenage years, although the patient is frequently unaware of the condition until it is fairly advanced. Deaf patients who develop retinitis pigmentosa often become aware of their loss of side vision sooner than normal-hearing persons who develop retinitis pigmentosa because they use their vision to com-

pensate for their poor hearing. They soon realize that they cannot participate in group discussions in which sign language is used. In such discussions they can see the person they are looking at, but they are unable to see other members of the group off to the side. For 85 to 90 percent of these people, loss of central vision is usually not a problem until later in life; however, the other 10 to 15 percent have a severe problem even during early adulthood. A person with a severe loss of side vision usually has less than 5 degrees of central vision and will have many problems using that vision.

Tests for retinitis pigmentosa. The following are some of the more common tests for retinitis pigmentosa:

1. One of the more well-known tests for retinitis pigmentosa consists of examining the inside of the eye at the fundus. In the fundus the choroid and its blood vessels are visible, giving the appearance of islands of pigment situated between the vessels. The fundus in the eyes of a patient with retinitis pigmentosa will show clumping of the pigment and narrowing of the blood vessels.

2. The dark adaptation test is conducted to measure the dimmest light that the patient can see after entering a dark room. Normally, a person experiences a rapid increase in his or her ability to see a dim light, which corresponds to an increase in the sensitivity of the retina or decrease in threshold over the first couple of minutes. With retinitis pigmentosa this adaptation is slow and incomplete because the rod system, which serves mainly side and night vision, is damaged.

3. In another test cone sensitivity is measured to obtain some idea of how well the cone system is functioning. A person may have 20/20 vision and still have trouble with his or her cones. In the standard test for visual acuity the contrast between black letters on white is so great that a person with only half of his or her cones could still have 20/20 vision as long as he or she has the proper refraction. Sensitivity (threshold) measurements can detect an abnormality which is often obscured by one's visual acuity. This type of specialized testing, however, would never be used in a screening program.

4. Visual field testing is used to detect the loss of side vision. With retinitis pigmentosa the loss of side vision typically begins in the midperiphery in a donut-shaped area of poor vision. This condition is called *ring scotoma.* In the more advanced stage, only the very central area of vision is left and, occasionally, a little peripheral island.

5. The electroretinogram is used to measure the normal electrical potential produced inside the eye in response to light. This machine is usually available only at large medical centers, although several companies are starting to manufacture portable units, which could be used in the doctor's office. However, an electroretinogram is usually not economically practical nor always necessary to make a diagnosis. Sometimes the machine is helpful in the prognosis because it can be used to establish the patient's variant or subtype of retinal degeneration. Sometimes it is helpful in reassuring the relatives that they do not have the disease. But the electroretinogram is more of a research tool which should be used on a referral basis rather than as a routine screening instrument for deaf-blind children.

6. The final test is called fluorescein angiography. This test involves the use of dyes which are photographed inside the eye. The dye shines through the areas where the pigment is missing. As the dye flows through the retinal blood vessels, it is visible and forms a different pattern from that formed by the dye showing through from the choroid.

Treatment of Retinal Damage

At this time no effective treatment is known for retinal damage. However, animal studies have shown that excessive amounts of light hasten retina damage in strains of rats and mice, which spontaneously develop genetic retinal degeneration. These studies compared the effects of light and dark on the eyes. The animals that were kept in the dark did better, which led to the idea of having patients wear an opaque contact lens over one eye for extended periods of time to protect it from the light. This theory is being tested at Harvard University, but researchers still do not know whether it is going to be useful. However, other evidence indicates that the best condition for the retina may be mild illumination during part of each day. Therefore, I think the best advice to give to the patient is to wear sunglasses outdoors and to avoid places such as the beach, where sunlight and glare are extreme.

No direct forms of treatment for retinal damage are available at the clinical level. I conducted a study with Vitamin A after reading in the literature that large doses of it may be helpful. However, I found that Vitamin A was helpful only in one very subform of retinal disease. Large doses of Vitamin A given over a period of a month did absolutely nothing for most of the patients with retinitis pigmentosa.

Genetic Considerations

The terms *recessive, dominant,* or *sex linked* are understood and used frequently today. But back in the days when Gregor Mendel, the Austrian botanist, was conducting breeding experiments with peas (1856-1870), and then later when his work was rediscovered by investigators during the so-called Cryptogenetics Era (1900), these terms were still a puzzle. At that time they did not know the mechanism of heredity, but they did know the mathematical ratios of inherited characteristics. When the chromosome theory evolved, it was determined that these mathematical ratios occur because genes are carried in packages—so many genes are located on one chromosome and a number of genes are situated on another chromosome. This theory explained why some genes seem to travel together, why different ratios occur in subsequent generations, and so forth. The theory also helped to explain why certain human traits and diseases occur in special patterns. Color blindness, for example, which should really be called color deficiency, is carried by the X chromosomes and is more frequent in males.

In the early nineteen hundreds ophthalmological syndromes were the latest thing in medical genetics. Since then much work has been done on bacteria and enzymes. The protein structures of many tissues are now known, and the mechanisms of structures at the cellular level have been established. The fields of ophthalmology and audiology, however, lag behind other fields of medicine in the genetic explanation of diseases because it is impossible to obtain useful biopsies of eye and ear tissues. Therefore, animal models must be used to obtain an understanding of the enzyme and protein structure. The reason this is important is that carrier detection often becomes feasible after the biochemical defect of a given disease is known. Occasionally, the discovery of a biochemical defect provides the breakthrough that leads to what is called rational therapy. Although it does not offer a specific cure, carrier detection can prevent some diseases because many genetic diseases are inherited in a recessive fashion. For example, if one in several thousand people is affected by a genetic disease, then maybe one in a hundred people who are perfectly normal is a carrier. So for every person who has the disease there are many people who are carriers. The disease cannot be disposed of just by genetic counseling of people who are affected by it. In fact, the only impact counseling has on many diseases is to lower the occurrence by 5 to 15 percent in the next generations, depending on the gene frequency in the population. Counseling will never get rid of diseases because most of the cases occur from other families that carry the gene and never come to light until there is an unlucky marriage of two carriers. In fact, good estimates indicate that most people carry about five to seven pathologic recessive genes. The reason most people have normal children is that they are lucky enough to marry someone who does not carry the same defective genes.

In summary, what I am suggesting is that more emphasis be put on programs that will promote research that detects carriers. When we have the capability to determine who is and who is not a carrier, then we can give good genetic advice to a lot of people.

Patient Counseling

What about the patient with Usher's syndrome who is in high school or college and who has been detected to be not only deaf but also suffering a progressive loss of vision? Certainly, something should be done about the victim's rehabilitation and education. I think it is important at this point that the patient be advised that he or she has a recessive disease. The patient must be made aware of the consequences of a marriage between two people with Usher's syndrome. The possibility of this happening is very real because deaf people often marry other deaf people. Their children, therefore, have a 100 percent chance of being affected by the disease. On the other hand, if a person with Usher's syndrome marries a person who is deaf from other causes, such as rubella, bilateral infections of the ears, or dominantly inherited deafness, then they have minimized the chance of their children being deaf and blind.

Rehabilitation and Screening Programs

People who are going blind do not want to learn braille until they are blind. However, deaf people who have overcome their handicap and who are faced with the loss of their vision seem to recognize a little sooner that they can make use of training. These people realize that they were able to function much better after they had learned the communication techniques available to the deaf. The patient will have lost two main means of communication if he or she waits until he or she is deaf and blind before starting rehabilitation training. However, I do not advocate locating these people when they are children in schools for the

deaf and telling them that they are going to have retinitis pigmentosa. I think these children do best in a school for the deaf as long as they have enough vision to get them through high school and maybe through college. At that time in life, however, many of them start to experience problems with mobility, reading speed, and communication in group discussions with sign language. Then it is realistic for them to know about their handicap and to start making plans for their occupational training. They are at an age when some information on the genetic aspects of the disease might have a bearing on whom they choose to marry.

I think, from a rehabilitation standpoint, that high school age is a good time to screen for retinitis pigmentosa. What is much better is more generalized screening in schools for the deaf for such things as poor visual acuity and increased intraocular pressure. Also, screening for hearing defects should be conducted in schools for the blind so that any form of disability is detected at an age when it is causing lack of function. I think screening programs should concentrate on detecting preventable eye diseases and defects.

Most of the patients with Usher's syndrome respond well at college age to a full explanation of their problem. The fact that the disease is slowly progressive in most cases gives the patient some latitude for coping with the shattering thoughts about the future. The patient's fears are usually counteracted by an awareness that his or her symptoms have been slowly accumulating and that proper training is available in centers for the deaf-blind. Such ambulatory training includes the proper use of the long cane, which the patient can utilize in situations where poor side or night vision is a problem. When reading becomes a problem for the patient, television image intensifiers, good lighting, and magnification should be tried before switching completely to braille or tactile signing. But perhaps the most important task is finding an occupational niche that the handicapped person can occupy.

Finally, we have found that some patients with Usher's syndrome are successful mothers. However, the patient's residual vision and the strong support of the patient's family have played important roles in these cases.

Examining the Deaf-Blind Child

Presented by Ted L. Johnston, M.D. Ophthalmologist
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Although a deaf-blind child may desperately need special education, his or her particular disability may not qualify him or her for participation in a federally funded deaf-blind program. Therefore, we attempted to determine which patients are legally qualified for this type of program. The following criteria are recommended as a minimum for certifying that a child is eligible for ESEA, Title VI-C, funding:

1. A child whose estimated corrected visual acuity is 20/100 or less, as determined by a fully trained ophthalmologist, and who has a minimum of 30 db bilateral sensory neural hearing loss, as determined by an audiologist who meets the minimum certification requirements of the American Speech and Hearing Association, may be automatically included in the deaf-blind educational program. The child will be subjected to continuous education evaluation during the program.
2. A child whose estimated corrected visual acuity is between 20/40 and 20/80 and who shows no evidence of using hearing for language development may be included in the deaf-blind program for a six-month evaluation period. At the end of the evaluation period, the ophthalmologist and audiologist who originated the evaluation placement and the educational diagnostician should certify that the child needs a deaf-blind program or he or she will be excluded.

In some programs it is not unusual to see a "blind" child stand up, walk across the room, and change the television channel to one that he or she would rather watch, or even adjust the set so that the picture is a little clearer. This child is not eligible by any definition to be taught in a deaf-blind program. But because of inadequate examination and evaluation of the child's visual function, he or she is occupying space needed by a truly deaf-blind child. We know that our examination of visual function, especially the subjective part, is not realistic in many of these cases.

When deaf-blind children are put into a good educational program, they begin to learn many skills. Many profoundly retarded children actually seem to learn to see when they are exposed to individual and well-directed teaching methods. They start to use their hands and eyes together. A rather typical example is a boy I had the chance to see several times in Omaha. This boy was a maternal rubella victim who had undergone surgery for cataracts. He was wearing corrective glasses. When the boy was first seen, he could find an object visually, but he would always look away and manipulate the object by tactile sensation. After two or three months in an intensive teaching program, he started using his hands and eyes together. He began to visually direct what he had previously been doing with his hands. This accomplishment was very exciting to me because the teacher and not the ophthalmologist was teaching the boy to see.

We do not really know how people learn to see. Most people have to learn to see within the first few months of their lives to have good vision. This emphasizes the terrible quandary that an ophthalmologist must be in when he or she encounters a rubella syndrome child who has either dense cataracts, an active inflammation, or an active viral infection of the eyes. This child is never going to have good vision unless he or she undergoes surgery at an early date. However, if the child is operated on too early, he or she may very well lose the eye from exacerbation of the infectious process. The job of the ophthalmologist and the optometrist is to estimate, and in many cases to guess, the visual potential of the child. The task not only requires interest and some ingenuity, but also those basic skills required in one's daily practice. We can say without hesitation that certain children are blind

and that other sensory modalities must be used. Some children have normal ocular equipment, but its value to them depends on other physical and mental factors. Some need treatment, lenses, or surgery, for example, which may determine the extent of their capability.

Examination Location

Where do we conduct our examinations? We usually see the child for the first time in the office, where the advantages are obvious. All of our comfortable equipment is available, and trained personnel are present to measure, test, and help. However, this location has some disadvantages. In our offices we do not have the time to sit down and calmly think about each child. We cannot investigate the child in terms of his or her degree of handicap. We have to guess more or less as to what will be useful to the child in the future.

We also do some examining in the hospital situation. But in my case that is usually done only when examination under anesthesia is necessary. An institutional setting is where I have done most of my work and where, in my opinion, the most valuable evaluation of a child's potential can be carried out. This evaluation is done best by a team approach. Our evaluation team includes a pediatrician, an orthopedist, an otolaryngologist, an audiologist, a psychiatrist, an educator with special training in the field, a physical therapist, a social worker who is familiar with the patient's family, and sometimes a nutrition expert. We have tried to pare down the number of disciplines involved but often find that we need the advice of the missing experts. All available information about the child is gathered in advance so that we are thoroughly familiar with his or her visual condition. Then we each report on the child and discuss diagnoses, recommendations, and probable progress.

Eye Screening Evaluations

My usual routine is to go into the ward with staff personnel who are interested in the children who are scheduled to be examined. In the ward I do a preliminary examination and obtain general physical information on the patient, study his or her behavior, and observe his or her function and, if indicated, tensions. In some cases, when the child is totally blind, this part of the original screening examination can be finished in 30 seconds. However, most of the children are given cycloplegic drops and another examination in about 30 minutes for retinoscopy and ophthalmoscopy.

Eye screening evaluations take much less time than most of the other testing modalities. I can screen 15 children while the other members of the evaluation team are working on one or two children. In this type of screening the children are rapidly divided into four main groups:

1. The blind children who do not see, who will not see, and who require other sensory input for training.
2. The children who have normal eyes with relative refractive error, no squint, and no need of ocular treatment.
3. The children who need treatment, glasses, surgery for cataract or squint, medication, or treatment for glaucoma. Firm recommendations need to be made for referral of these children to some place where the indicated treatment can be obtained. These recommendations must be made in reasonable relationship to the child's overall condition.
4. The children who need referral for examination under better conditions with more sophisticated instrumentation. This group includes children whose cooperation is not good enough to permit the examiner to conduct a comprehensive examination.

Most doctors donate time from their practices for worthwhile programs. I think that an occasional day spent in a screening activity of this type is very worthwhile. An examination session at an institution improves your recording speed, your clarity of diagnosis, and your specificity of recommendations. This examining location provides an extremely economical way to see a lot of children in a reasonable amount of time. The program or institution can usually find the funding to pay something for your office overhead.

I spent two days in Beatrice, Nebraska, at the state school, where I screened 61 children. No recommendations were made for 49 of the 61 children because they either had no eye condition that needed treatment, their eyes were incapable of improving, or their eyes were normal to examination. Of the 61 children examined, 3 had cataracts, 6 had retinitis pigmentosa, 10 were totally blind, 23 had exophoria, and 16 were normal. In addition, 5 children were recommended for examination under anesthetic, 5 were recommended to wear glasses, and 5 were recommended for a special teaching routine. Some of the children listed in this partial summary of findings were included in more than one of the categories.

By 4:30 p.m. on the second day of the tour I had looked at the last child. I sank onto a bench

and said, "God, I'm tired." Much to my surprise the two young people who had assisted me also indicated that they were tired. "But of course they are tired," I thought. "They had carried the children, held them in the chair, and held them on the floor. They had done most of the hard physical labor." Staff personnel and I felt that the two days were very worthwhile because we now had some idea of the state of the children's eyes, and their teaching programs and care could be related to visual reality.

Examination Instruments

The most important piece of equipment to take along for this kind of work is jeans, because you will spend a lot of time on the floor. Every institution has a room that can be darkened, even if it is just a broom closet. Sometimes you have to move the brooms and pails out of the closet, but it is still a place where you can take a child and look into his or her eyes with a retinoscope or an ophthalmoscope.

I also take direct and indirect ophthalmoscopes and retinoscopes, each with its own transformer, a penlight, Kleenex, ophthane medriacyline (½ percent), thermometer, alcohol, sponges, and an old red parka. I usually carry a small cassette dictator. When I finish an examination, I dictate my report right at the examination location rather than depending on my memory for preparing the report at a later time.

You must also have someone from the institution staff accompany you during the entire tour. This person should help you with the patients, help you keep records, and do whatever else is necessary.

Conclusions

Although a day or two spent in a screening activity may not be rewarding financially, it is good exercise for the mind and body. You may pick up some interesting pieces of information for your own practice, especially if you are working in your specialty.

It is impossible to categorize deaf-blind children and to say that there is a technique that will work for all of them. Just as you work differently with each patient that comes in your office, you will work differently with each deaf-blind child in an institution. You will modify your examination techniques and your approach with each patient, which is something that no one can teach you—it must be learned through experience! You can make progress with these children by physically touching and keeping constant contact with them while you are doing your examination. However, you have to use your own experience and your own routine examining procedures. These procedures are the ones you know best and the ones that will give you information on the patient's eyes.

Assessing the Deaf-Blind Child

Presented by Laurice Jenkins
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At the California School for the Blind, we provide in-depth educational evaluation of the deaf-blind child, including those psychological, audiological, and ophthalmological aspects of the child's functioning which have a direct bearing on his or her education. Specifically, what we do is try to determine the best methods and procedures for working with the child and for ascertaining his or her potential. Sometimes this is quite possible, and sometimes it is not.

Most of the population at the school is multihandicapped. Even if these children were not blind or deaf, they still would not be able to adjust to a normal school situation. More than likely they would be in some kind of program for the multihandicapped or at least a program for the educationally handicapped.

We try to maintain a flexible program for the child because he or she has to adapt to so much. Each child is allowed as much time as possible to settle into our program and become acquainted with us. Some children never do adapt and never will, but not too many. We have had a few children who were just too disturbed and who did not respond to any situation. However, most children are reasonably happy in the evaluation situation within one or two weeks.

Insofar as possible we try to answer specific questions asked by teachers, parents, and other people involved with these children. We also make recommendations on the placement of a child by level or by type of program. We do not recommend specific programs because they can vary, and we are not familiar with all programs. We recommend, for example, a deaf-blind day program rather than the deaf-blind day program of the Smith Unified School District. We try to be nonspecific to avoid the problems so often involved in recommending specific programs.

Enrollment Procedure

The preliminary enrollment procedure is very much like that at any other school with full-time students. First, we receive a referral from the regional deaf-blind center for a child who has either come to their attention or our attention. Then we write for medical, social, and educational information. After we have received this information, the parents are contacted to set a meeting date that is agreeable to them. We do not send out a form that assigns the parents a specific date and hour to appear at the school. Instead, we work with them individually and try to accommodate our admission procedures to the parent and to the school situation. When the child comes for evaluation, we do require that a parent accompany him or her during the first week. However, we often make exceptions because the parent or adult most familiar with the child may not be able to make the trip. In that case the child is given accommodations in our dormitory. If a parent, older sibling, or grandparent can come with the child, we provide room and board for them at no cost. They have to arrange for their own transportation, however. Sometimes their expenses are paid by the district. Having a parent in attendance helps to get additional information about the child and helps the child to adjust to a strange environment. The parents also feel more comfortable about leaving their child with us for the remaining three weeks of the evaluation period.

Evaluation Period

During the first week, we do very little work with the children because the environment is strange for most of them, except those coming from an institution. We let each child find his or her own way around and become accustomed to us. If the parent is available during the first week,

we do try to perform an audiological evaluation with the parent in attendance. Otherwise, we usually defer this evaluation until the second week. The child is also checked by our ophthalmologist during the first week. The ophthalmologist does not give the child a complete examination, but the child can be checked to determine whether further examination is needed. Occasionally, the ophthalmologist will refer the child for additional testing. Most of our children are rubella syndrome victims who had their visual problem identified at an early age. Their hearing problem is discovered later. Sometimes the child reaches school age before the parents are really aware that he or she has a hearing problem, particularly if it is not severe and the child does respond to some sound. During the remainder of the three weeks we work with the child primarily in the classroom. The psychologist becomes involved during the third week, after the child has had a chance to settle down and become acquainted with everyone. In this way we can obtain a more valid evaluation of the child.

At the end of the evaluation period, the last day normally, the professionals who were involved with the child are invited to a staff conference. At this meeting we discuss the child and make recommendations, which we try to make cooperatively in terms of what is feasible and what is available, not what is ideal. Within two or three weeks, we issue a final staff report, which is sent to all the professionals involved. A copy of the recommendations for educational programming (the longest section in the report) is sent to the parents to provide them with feedback from the evaluation. We also send out a short questionnaire to determine whether the parents and the professionals who were involved with the case feel that our services have been of help to them. The comments we receive back on the questionnaires are sometimes interesting, particularly those from the professionals who can be more critical than the parents.

In several instances we had children referred for reevaluation after two or three years. This happens when the child's progress does not follow that previously predicted for him or her, or when a great deal of change has occurred in the child, or when the child has undergone virtually no change. People are now becoming aware that reevaluation is possible.

Entrance Criteria

To enter the assessment center, the child must meet certain criteria. He or she must walk reasonably well. We do struggle along with children who walk with difficulty as long as they have the endurance to walk distances. Sometimes we receive undue credit for physical therapy—children who can barely walk when they first arrive are often walking quite well at the end of four weeks. The layout of the school requires that the children do a lot of walking.

We do not have any age limit, but we have to be careful with children who are very young or very large. The very small child may get trampled or the very large child may hurt other children, so we have to take these factors into consideration. However, this does not mean that we do not take these children. We even take nonambulatory children if someone provides an attendant to wheel the child back and forth in a wheelchair. A very young child can be bussed in and out during the day and spend a minimum of time in the dormitory.

We also take children who require regular medication for such things as diabetes and behavior control. If a child is on behavior control medication, we try to get him or her off the medication with the consent of the parent, the child's physician, or our own physician and staff. We have taken many children off medication for behavior control only to find that they have become better behaved and better controlled. I do not say that we never use sedatives or medication, but we use them very carefully and only after we have tried the children without it.

Children who are not toilet trained, who need help with feeding, or who have severe emotional problems are also accepted at the school.

Visual Acuity Assessment

I will not go into the mechanics of assessment, except those for visual acuity. If a child has some light perception, we try to determine how much. We find out whether the child wears glasses or has glasses available. Sometimes a child has glasses but does not wear them. If so, the parents are requested to bring the child's glasses so that our ophthalmologist can check them. The following observations are made by our staff and are reported and correlated with the ophthalmologist to obtain some idea of the child's visual functioning:

1. Can the child detect an M&M candy or a shiny toy?
2. Does the child reach beyond or short of the object?

3. Does the child hold the object up close to his or her eyes to inspect it, or does he or she just make a passive examination?
4. Does the child respond to light or motion and track a light source or light shown on the wall?
5. Does the child respond to light motion from the side?
6. Does the child try to see objects on the side or give any indication of seeing something on the side?
7. Is the child afraid to walk down steps?

These tests are made both formally and informally. If the child can see an object and reaches for it, we know that he or she at least has some sight. We then measure the distance in terms of actual feet that the child can detect an object, and we record it in the report. We do not attempt to work out the ratios, but we do get a reasonably accurate picture of the child's visual acuity. Although these are not formal scientific tests, they can be helpful to a teacher who may not have thought of checking the child's vision in this manner.

Then we report the findings to the teacher and suggest how he or she can help the child with additional work. If the doctor uses terminology that we do not understand, we ask him or her for clarification. We always try to secure or report our information in terms that are clear and can be understood by the teachers and parents.

Hints for Examining and Reporting the Vision of Deaf-Blind Children

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Because of his or her unique combination of handicaps, the deaf-blind child may offer quite a challenge to the examiner during a visual examination. A typical rubella patient is mentally retarded and physically underdeveloped. He or she has congenital cataracts, severe nerve hearing loss, and sometimes systemic problems such as a heart defect. Most of these children have no vocal language and adjust poorly to unfamiliar surroundings and to doctors who probe around their eyes. Although these children may be mentally retarded, they often outsmart doctors, so that they must be examined under anesthesia. An understanding of the deaf-blind child's likes and dislikes, temperament, and growth and development pattern may help the examiner to establish a good rapport with the patient and to make the experience more pleasant for both.

Examination Conditions

The examiner should remember that the deaf-blind child usually adjusts poorly to a new environment and to new people. In particular, the deaf-blind child finds it difficult to adapt to the examination chair. Therefore, the examiner must adjust his or her techniques and instruments so that they may be used in the deaf-blind child's unique situation. Many examiners, for example, work with the child on a rug on the floor.

An important thing to remember is to allow sufficient time for a visual examination. This time is needed to establish a rapport as well as to perform the actual examination. If the doctor cannot afford to spend this much time, he or she should not accept deaf-blind children as patients.

No two deaf-blind patients are alike, and each may require a different approach for a successful examination. A parent and a teacher should be present so that a complete case history can be obtained. A child often exhibits certain skills at school that he or she does not exhibit at home and exhibits certain behaviors at home that he or she does not exhibit at school. Some of the things that can be learned about the child's visual skills during the history investigation are whether the child sees details at a distance and at varying near-points and whether he or she uses vision in everyday activities such as eating and working.

When the patient first appears in the examination room with his or her parents or teacher, he or she should be allowed to explore any part of the room. The examiner usually sits in the middle of the floor, where the patient can walk up and reach out for him or her. The examiner should give tactile attention to the patient by patting, gently stroking, or tickling, depending on the patient. When properly performed, this tactile sensory experience is appreciated by the deaf-blind child, and the child will usually stay with the examiner for further testing.

A penlight is introduced after a good rapport has been established with the child. If the child does not enjoy the penlight, a favorite toy is placed within his or her reach. However, most rubella children love light; in fact, deaf-blind children usually are attracted to the overhead lights in the classroom.

When a deaf-blind child is examined, the examiner should use an indirect light, such as a penlight, in the room so that the child is not distracted by the ceiling lights. After the penlight has been introduced, the child will usually look directly at it, and the examiner may then proceed with recording the patient's response or pupillary reaction. Oculomotor skills may also be determined by the use of the penlight.

Lights will often cause a behavior change in the patient, which should be determined and explained in the visual report. The examiner should refer to the child's case history to make certain that he or

she is not subject to seizures. A blinking light may trigger seizures in certain children. Gross visual field studies may be performed by using a pair of penlights. Occasionally, colored filters are used to observe the patient's preference for colored lights versus white lights. One color may soothe a child, while another may excite the same child. This information is used in the examination and, many times, part of the examination is conducted with the patient looking through a colored filter.

The rubella child's familiarity with light is used in performing ophthalmoscopy and retinoscopy. The ophthalmoscope is introduced like a penlight while the child is lying on the floor, except that the examiner looks through the instrument. The patient, who is attracted to the light, usually reaches for it and pulls it toward his or her eyes with the examiner at the other end. The media and macula area may be examined in this manner, but an assistant is needed to hold a penlight off at the proper angle for examining the optic disk and other areas of the eye. If properly performed, this technique allows for an examination that is less frustrating for the patient and for the doctor. Retinoscopy examinations are performed in a similar manner. The child again is in a prone position on the floor, and the examiner holds trial lenses while using the retinoscope at an appropriate distance. If the patient is aphasiac, the examiner does not have to be concerned with providing the patient with any accommodations.

Measurement of Visual Acuity

The measurement of visual acuity may be frustrating for the examiner if he or she is not prepared to conduct the examination. Few deaf-blind children can be tested with a letter chart. Picture cards of various sizes may be matched by a nonverbal patient if he or she is functioning on an appropriate level. The use of toys or favorite objects placed at various distances from the patient will give an approximation of visual acuity for lower functioning children. Teachers may be of invaluable assistance in securing valid and reliable visual acuity ratings. A deaf-blind child, particularly if he or she is a low functioning one, may demonstrate more efficient vision from one day to the next day. A properly trained teacher can obtain an accurate visual acuity rating by placing objects that attract the deaf-blind child at various distances over a period of days, weeks, or even months. The teacher may also use lights and objects over a period of time to determine the deaf-blind child's usable field of vision.

The visual field is extremely important because many severely handicapped children have retinal damage and may use only certain areas of their retina for vision. A teacher needs to identify these areas because he or she can only be effective if material is presented in the usable part of the child's field of vision. Therefore, objective measurements of the patient's visual field and visual acuity are desirable. The use of an optokinetic drum is beneficial in determining whether form perception is present. Also, the visually evoked response instrumentation is valuable for objective measurement of visual acuity, color vision, and refraction.

Determining Level of Function

The examiner can learn much by observing the child in both a structured and an unstructured environment. The child's functional vision skills may be observed while he or she is exploring the examining room. The examiner should observe whether the child uses only vision skills in moving around or whether the child feels for things. The presence of eye contact should also be determined. Eye-to-eye contact is essential for teaching the child visual language and social skills. Eye-object contact is also important because many low functioning children do not look at the objects that they manipulate.

The visual examination should include not only the sensory aspect of vision but also the motor and perceptual aspects, because they are equally important in the visual performance of the deaf-blind patient. Although visual acuity cannot be taught, motor and perception skills can, and they follow a developmental sequence. The child's present level of function in all areas of vision must be determined, because this information is needed by the teacher to plan an educational prescription.

Examination Report

The examination report should include for the teacher, in addition to a structured eye information sheet, a narrative in layperson's language on the observations of the patient's vision functioning and information obtained from the case history.

When making recommendations for an educational program, one must remember that a single examination does not always provide a true picture of the patient's visual functioning, and care should be exercised in making important decisions. Many times a staff meeting in which all the members give their impressions of the patient is the most worthwhile. Greater insight may result from shar-

ing observations of the child, and a more meaningful report may be written.

The teacher may be helpful in securing additional information about a deaf-blind child's visual functioning. In the teaching process, visual acuity and visual field measurements can be made in a reliable manner if the teacher has been properly trained. Teamwork is essential for a meaningful visual examination. Teachers may be the source of various information, and they should receive reliable information. Examples of questions that would provide useful information for the teacher are as follows:

1. What is the cause of the visual impairment?
2. Is any special treatment required? If so, what is the general nature of the treatment?
3. Is the visual impairment likely to get worse, better, or stay the same?
4. Should the teacher be alert to any particular symptoms (such as eye rubbing and so forth) that would signal the need for professional attention?
5. What restrictions should be placed on the child's activities?
6. Should the child wear glasses or contact lens? If so, under what circumstances?
7. Were you able to make an accurate measurement of visual acuity? If so, what was the child's visual acuity?
8. If a visual acuity measurement was not possible, what is your opinion regarding the child's visual functioning?
9. Are the child's focusing ability and eye muscle balance adequate? If not, please describe.
10. Were you able to determine the field of vision? If so, were there areas of no vision in the field? Where?
11. Was the child able to follow visually a moving object? Were there directions in which he or she could not track moving objects? Which directions?
12. Will the child work better with large or small objects and pictures? At what distances?
13. What lighting conditions would be optimal for his or her visual functioning?
14. What are your specific recommendations concerning this child's use of vision in learning situations?
15. When should this child be examined again?

In conclusion, the visual practitioner is a member of an interdisciplinary evaluation team of deaf-blind children, and he or she should be aware that teamwork is an essential element for the success of the evaluation.

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